

DEVELOPMENT FEATURES OF FAMILY, CONGENITAL GLAUCOMA

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Abstract. *This article is devoted to studying the features of the development of congenital glaucoma in children with familial glaucoma. The authors examined two families with familial, congenital glaucoma. The patients were treated as inpatients in the eye department of the clinic of the Tashkent Pediatric Medical Institute.*

Keywords: *congenital glaucoma; primary congenital glaucoma; ophthalmoscopic examinations; keratometry; visometry; biomicroscopy; retina.*

Actuality. Glaucoma is a group of eye diseases manifested by a constant or periodic increase in intraocular pressure due to impaired outflow of aqueous humor from the eye, which causes depression of vision and atrophy of the optic nerve [2,3,4,9,11]. Glaucoma is one of the most serious diseases in ophthalmology, especially in pediatrics, and occupies a leading place among the causes of blindness and low vision [1,5]. In the literature, glaucoma occurs in about 3% of the world's population (about 70 million people). Researchers indicate that only half of all patients receive adequate treatment.

Less than 7 million patients suffer from blindness in both eyes due to glaucoma, and their number is steadily increasing [6,7,8,10,12,13]. The presented statistical data indicate that today there are still difficulties associated with both the diagnosis and treatment of this disease [15,17].

WHO believes that there are 70-100 million patients with glaucoma, and in ten years this number will increase by 10 million; in the world, 1 person goes blind every minute from glaucoma and 1 child every 10 minutes? It is believed that by 2030 the number of patients with glaucoma will double.

Patients with newly diagnosed glaucoma have many advanced stages. In Europe there are 160 thousand people blind from glaucoma, with 6.7% in Denmark and 20% in Sweden. Glaucoma is in second place after cataracts and first in disability among permanent vision loss [1,4,14,18,19,20].

According to the classification, congenital glaucoma is divided into hereditary, familial and sporadic categories. Hereditary glaucoma is defined when more than 3 relatives, including the proband (index case), have glaucoma in 2 consecutive generations, one of whom must be a first-degree relative of the other two. Familial glaucoma includes more than 2 first and/or second-degree relatives and does not meet the criteria for hereditary glaucoma. Sporadic glaucoma affects one patient with no first- or second-degree relatives affected.

Genetic studies have shown that more than 50 percent of glaucoma cases are familial. The incidence of glaucoma can be 10 times higher among people who have siblings with glaucoma. The three classes of glaucoma differ not only in the nature of inheritance and the degree of familial aggregation, but also in the clinical picture, course and severity of the disease.

The purpose of the study was to study the features of the development of congenital glaucoma in children with familial glaucoma.

Materials and methods of research. During the period 2020-2022, we examined two families with familial, congenital glaucoma. The patients were treated as inpatients in the eye department of the clinic of the Tashkent Pediatric Medical Institute. All patients underwent ophthalmological (visometry, biomicroscopy, A/V scanning, keratometry, ophthalmoscopy), clinical and laboratory research methods, consultations with related specialists (ENT, pediatrician, anesthesiologist, cardiac surgeon, neurologist).

When performing ophthalmoscopy, it is necessary to pay attention to the following parameters: size and shape of the optic disc, size, shape, degree of blanching, (pink or decolorized) neuroretinal rim; the size of the excavation relative to the size of the optic disc, the configuration (the nature of the temporal edge: flat, steep, undermined) and the depth of the excavation (shallow, medium, deep); indicator of the ratio of the maximum excavation size to the diameter of the disc of the optic disc (E/D); severity of peripapillary choretinal atrophy; the location of the vascular bundle and the associated symptoms of “puncture” and desolation of the optic disc encircling vessel; hemorrhages on the optic disc; the diameter of the retinal arterioles and the condition of the retinal nerve fiber layer.

Also, all patients had a carefully collected medical and life history.

Research results. In the first family the marriage is not related. In 2010, a boy was born with primary congenital glaucoma. The child was diagnosed by a neonatologist during an examination of the child in the maternity hospital. At birth, an increase in the eyeball by 3.0 mm was detected in comparison with the age norm. There was an increase in corneal diameter and severe keratopathy. Over the course of 13 years, the child underwent surgery three times in each eye. After surgical interventions, no stage of compensation from the organ of vision was observed.

The patient was prescribed continuous instillation of eye drops that reduce intraocular pressure to stabilize the process. In 2024, twins were born in this family, girls, who were diagnosed in the maternity hospital: OU - primary congenital glaucoma, advanced stage. Since girls showed an increase in the eyeball by 4.0 mm depending on the age norm, severe glaucomatous keratopathy was also observed. In which examination of the eyeball by ophthalmoscopy did not allow viewing the underlying structures of the eye. The girls were examined in a hospital setting and antiglaucomatous surgery was performed. The older brother also underwent repeated surgical treatment using biodegradable drainage GLAUTEX. All three children from the same family were discharged home in satisfactory condition.

In the second family in 2016, a boy was born with a diagnosis of OU - primary congenital glaucoma, advanced stage. Over the course of 10 years, antiglaucomatous surgery was performed four times on the right eye, and once on the left eye. In 2022, a girl was born in this family with a diagnosis of OU - primary congenital glaucoma, terminal stage, buphthalmos. The diagnosis was made in the maternity hospital. The patient with the above diagnosis, in order to exclude the diagnosis of retinoblastoma, was referred and examined by an onco-ophthalmologist, Doppler ultrasound of the organ of vision was performed, and therefore oncopathology was excluded. After which she was hospitalized in the eye department of the clinic of the Tashkent Pediatric Medical Institute. The child underwent antiglaucomatous surgery and was discharged home in satisfactory condition with a recommendation to be monitored by an ophthalmologist at his place of residence.

Conclusion. Thus, an analysis of the clinical and functional state of the eyeball in children with congenital, familial glaucoma showed that in these patients the manifestation of congenital glaucoma with an increase in the anteroposterior size of the eyeball and the diameter of the cornea,

severe keratopathy manifests itself immediately after birth. A preliminary diagnosis is made immediately after the birth of the child in the maternity hospital by a neonatologist. According to the analysis of the course, familial congenital glaucoma is refractory and difficult to treat, which is why these patients who have previously undergone repeated antiglaucomatous surgical interventions need to undergo sinustrabeculectomy with the use of drainage surgery for repeated operations. The use of drainage surgery in the above cases is considered one of the main ways to reduce and maintain ophthalmotonus in the surgical treatment of refractory glaucoma.

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